

ARTHROPATHY

I. INFECTIVE

A. Acute:

- Bacterial: "Septic arthritis"
 - Gonococcal, Meningococcal.
 - Brucellosis, Bacillary dysentery.
 - Staph, Strept.
- Viral:
 - IMN, HBV, HIV.

B. Chronic:

- TB.
- Syphilis.
- Lyme disease: *fever, erythematous rash, acute arthritis*, due to infection by a spirochaete.

II. IMMUNE

A. Acute:

- Rheumatic fever.
- Rheumatoid arthritis.
- Reactive arthropathy: *arthritis following intestinal infections*, e.g. Salmonella.
- SLE.

B. Chronic:

- All collagen diseases: RA, SLE, Scleroderma, PAN.

III. DEGENERATIVE

- Osteoarthritis.

IV. METABOLIC

A. Acute:

- Acute gouty arthritis.

B. Chronic:

- Chronic gouty arthritis.
- Hemochromatosis & amyloidosis.

VISIT...

LANZAROTE
Caliente.COM

V. MISCELLANEOUS

A. Acute:

- Traumatic arthritis.
- Hemarthrosis (Hemophilia).
- Henoch-Schonlein purpura.
- FMF.

B. Chronic:

- Sarcoidosis.
- Charcot's joint (Neuropathic joint).
- Congenital syndromes with arthropathy:
 - Ehlers-Danlos syndrome.
 - Marfan's syndrome.
 - Osteogenesis imperfecta.

Ehlers-Danlos Syndrome

Genetic weakness of the CT:

- **Blood:** vascular purpura due to vessel wall defect.
- **Joints:** hypermobile joints due to weak ligaments.

Marfan's Syndrome

- **Tall stature.**
- **High arched palate.**
- **Arachnodactyly.**
- **Joints:** dislocation of the joints (loose joints).
- **Eyes:** dislocation of the lens.
- **Heart:** aortic regurge, mitral regurge.

Osteogenesis Imperfecta

- **Bones:** fragile bones with repeated fractures & deformities.
- **Joints:** hypermobile joints with degeneration.
- **Eyes:** blue sclera.
- **Ears:** early deafness.
- **Stunted growth.**

ترجع مرة بعد ٨-١٠ أسابيع مع الارتفاع المتكرر بعد ٨-١٠ أيام

unknown (not auto imm)

noninfective

oral

3

Familial Mediterranean Fever

Incidence: in Jews & Arabs, in childhood & adolescence.
Presentation: attacks lasting for few days & recurring every few weeks or Months
Attack: FUO fever, severe abdominal pain (peritonitis), chest pain (pleurisy), or (pericarditis), or effusion, & arthritis.
Complications: some cases may develop amyloidosis, Nephrotic S & renal failure.
TTT: colchicine to prevent & cure the attack.

no investig

DD of Polyserositis

chronic infectn
chronic inflam

Septic (Suppurative) Arthritis

Etiology: Staph, Strept, gonococci, meningococci.
Clinical picture: Severe monoarthritis (usually of a big joint), fever.
Synovial fluid: Turbid with high number of PNLs.
TTT: Antibiotics + drainage of the synovial fluid.

Charcot's (Neuropathic) Joint

Definition: joints damaged by trauma due to loss of protective pain sensation.
Etiology: PN especially DM, Syringomyelia, Tabes dorsalis of syphilis.
Clinical Picture: Swollen painless joints, + abnormal range of joint movement.
TTT: ttt of the cause, + surgery in advanced cases.

proprioceptive

OSTEOARTHRISIS

خشونة مفاصل
حاجات صغرى

DEFINITION

- Degeneration of the joint cartilage, with proliferation of bone & osteophyte formation.

ETIOLOGY

- Primary:** (hereditary)
 - Idiopathic:** there is hereditary predisposition.
 - Incidence:** common in old age especially in obese females.
- Secondary:**
 - Obesity,** Deformity, Damage by trauma, Endocrinal diseases (e.g. acromegaly), Gout.

A → chronic

White Knight Love

CLINICAL PICTURE

1. Site:

- Weight-bearing joints: knees, hips, cervical & lumbar spine.
- In primary OA: the terminal IP joints are affected.
- In secondary OA: other joints are affected according to the cause.

2. Manifestations:

- Main symptom: joint pain & stiffness which ↑ by movement & ↓ by rest.
- Spine affection: manifestations of spondylosis (refer to neurology).
- Main signs: joint crepitus, tenderness, ↓ range of movement, & lately: muscle weakness.
- Herberden's nodes: these are pathognomonic & develop on the dorsal aspect of the distal IP joints.
- Bouchard's nodes: these are similar nodes on the proximal IP joints.

INVESTIGATIONS

1. Laboratory:

- Normal ESR, Normal CRP, Normal uric acid.
- Negative RF, Negative ANA.

2. X-ray:

- Narrowing of the joint space. bone proliferation
- Osteophytes (lipping).

TREATMENT

1. General measures:

- Weight reduction in the obese, physiotherapy.

2. Medical TTT:

- NSAIDs.

3. Surgical TTT:

- Arthrodesis, Arthroplasty. joint replacement

- Gout: metabolic disease : $\uparrow$ uric acid
- Hyperuricemia: investigational (clinical presentation)

GOUT

DEFINITION

- Abnormality of uric acid-metabolism that results in the deposition of sodium urate crystals in:

- Joints, leading to **gouty arthritis**.
- Kidneys, leading to urate stones & **gouty nephropathy**.
- Skin, leading to **gouty tophi**. (soft tissue swellings s.c. nodules)

clinical presentation

ETIOLOGY

I. RENAL GOUT

90% of the cases

- It is due to decreased excretion of uric acid.

1. Primary: "The most common"

- It may have a genetic origin & is more common in males.

2. Secondary:

Acute & chronic renal failure.

- Acidosis: ketoacidosis & lacticacidosis.

- Dehydration. $\downarrow$ GFR $\rightarrow$ $\downarrow$ excretion

- Drugs: **diuretics** (thiazides, frusemide), **excess alcohol**, **low dose aspirin**.

Directly: $\downarrow$ filtered load of excess U.A.

GRF

U.A. needs alkaline medium to be excreted

to buffer acidosis $\rightarrow$ kidney: treats. of HCO_3^- so, $\downarrow$ HCO_3^- in urine $\rightarrow$ acidic urine $\rightarrow$ U.A. excretion

Probenecid
pyrazinamide
anti TB

II. METABOLIC GOUT

10% of the cases

- It is due to increased production of uric acid.

1. Primary: "The most common"

- It may have a genetic origin & is more common in males.

2. Secondary:

a) Enzyme defects:

"Lesch-Nyhan syndrome"

- Deficiency of the enzyme: Hypoxanthine-guanine transferase.
- X-linked disorder with: gout, mental retardation, choreo-athetosis.

b) Increased turnover of Purines:

- Myeloproliferative disorders: e.g. PRV & CML especially with $\uparrow$ $\uparrow$ $\uparrow$
- Carcinoma.
- Psoriasis.

inherited on sex chromosomes
also hemophilia
G6PD def.
Duchenne
 $\downarrow$
 δ sufferer
 ϕ carrier

ribonucleotides (nucleic acids) $\rightarrow$ Purines $\rightarrow$ Xanthine $\rightarrow$ Hypoxanthine $\rightarrow$ Guanine
HGT
Xanthine oxidase
U.A.

shift on side of U.A. pathway
 $\downarrow$
B.G.
 $\uparrow$ U.A.

chemotherapy
 $\uparrow$ destruction of cell
Nucleic Acid
"Tumor Lysis Syndrome"
White Knight Love

* Symptoms → level just below the knee
 * asyp → pain

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CLINICAL PICTURE

I. ASYMPTOMATIC HYPERURICEMIA

II. ACUTE GOUTY ARTHRITIS

D.D. of Acute Arthritis

- Precipitating factors of joint affection:

- Excessive ingestion of purines, excess alcohol, diuretics, infection. (turn over gphn [tissue breakdown])

- Distribution of joint affection:

- Number:** Monoarticular: in most of the cases, Oligoarticular: sometimes. < 4 joints (2 or 3)

- Site:** Metatarso-phalangeal joint of the big toe in 50 % of the cases. Most Common (MCQ)
- Other affected joints: Ankles, heels, knees, wrists, fingers, elbows.
- Usually spared joints: Hips & shoulders.

- Manifestations of joint affection:

- Acute onset of: severe pain, tenderness, swelling, hotness, redness + limitation of movement.
- Fever: and general constitutional symptoms.

- Course of joint affection:

- Attacks may be repeated & the condition may pass into the chronic stage. (polyarthritides)

III. CHRONIC TOPHACEOUS GOUT: & Arthritis

1. Chronic arthritis:

- Joint affection is:** Polyarticular, asymmetrical.
- Joints are:** chronically inflamed & the condition ends in secondary osteoarthritis.

2. Tophi:

- They are:** skin nodules due to urate deposition around joints & cartilage (ear pinna).
- They are:** yellowish white nodules that may ulcerate & discharge chalky material.

IV. GOUTY NEPHROPATHY: → Mortality (morbidity & chronic)

1. Urate stones:

ARF

leading to obstructive uropathy. (Radio-lucent)

2. Chronic interstitial nephritis.

3. Chronic renal failure.

4. Acute urate nephropathy:

- It occurs: especially in patients with Myeloproliferative disorders: e.g. PRV & CML especially with ttt.
- It leads to: acute tubular obstruction by urate crystals, leading to acute renal failure.

ARF → Sudden massive
 CRF → I.V. fluids

ICRF

HTN
 LDM
 Gout

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INVESTIGATIONS

1. Serum uric acid:

- Elevated (normally: 2–7 mg %).

2. Investigations of the cause:

- e.g. CRF, CML, myelocytes (B.M biopsy) & urea & creatinine (RFT)

3. Investigations for the joints:

- X-ray: soft tissue swelling, bone erosion.
- Aspiration of synovial fluid: urate crystals.

4. Investigations for the kidneys:

- Imaging: X-ray (stones), IVP (stones & renal function), Ultrasonography (stones).
- Renal function tests: urea, creatinine, creatinine clearance → early of ↓ GFR

5. Biopsy from the tophi.

Na urate crystals & synovial granuloma.

DD from other causes of acute & chronic arthropathy (itis)

TREATMENT

A) TTT of Acute Gouty Arthritis:

1. Bed rest.

2. Excessive fluid intake.

3. Anti-inflammatory agents:

i. NSAIDs

Colchicine:

- 1 mg / 2 hrs orally until symptoms disappear or diarrhoea appears.

ii. NSAIDs:

- Indomethacin: 50 mg tds.

iii. Corticosteroids:

- Indication: when there is contraindication or no response to [colchicine or NSAIDs]
- Dose: Prednisone, 1 mg / Kg / day orally.
Intra-articular corticosteroids may be given in severe cases.

4. Allopurinol & uricosuric drugs: are not used in ttt of Acute Gouty Arthritis.

as may worsen condition by initial unexplained ↑ U.A.

Q: Acute gouty

+ Chronic & if Q chronic acute

Uses of Colchicine

- 1- PMF
- 2- Gout
- 3- Behcet
- 4- ankylosis (antifibrotic)

لازم توقف و تنقل و Cortisol

drugs w ↑ liver enz:

Oral

statin
Amiodarone

[Rifampin
INH

[All hepatotoxic drugs
Allopurinol

chronic arthritis
↳ tophi skin

B) TTT of Chronic Tophaceous Gout & Gouty Nephropathy:

1. Anti-hyperuricemic drugs:

a) Allopurinol:

"decreases the synthesis of uric acid"

Zyloric

- Action: inhibits the enzyme Xanthine oxidase essential for conversion of xanthine to uric acid.
- Dose: 300 - 800 mg / day orally.
- Side effects: dermatitis, diarrhoea, elevation of liver enzymes.

فشل العلاج

b) Uricosuric drugs:

"increase uric acid excretion by the kidney"

يزيد إفراز

- Probenicid: 250 mg twice / day.
- Sulphinpyrazone: 400 mg / day.

These drugs should be given with plenty of fluids + alkalinization of urine to avoid nephrolithiasis

washout

Side effect: transient early unexplained hyperuricemia
للوقت

2. Symptomatic TTT:

a) Surgical removal of tophi.

b) TTT of renal stones.

c) TTT of acute urate nephropathy:

- Allopurinol.
- Excessive fluid intake.
- Alkalinization of urine: by sodium bicarbonate.

by NaHCO₃
as U.A. dissolver (increases)
in alk. medium

d) Anti-inflammatory drugs for chronic arthritis: e.g. NSAIDs.

Colchicine only in acute cases (due to toxicity)

C) TTT of asymptomatic hyperuricemia:

- TTT with Allopurinol is indicated only in:

- Family history of gout.
- Serum uric acid: more than 11 mg %.

(↑ incidence of complications)

(↑ incidence of complications)

TREATMENT

1. Prednisone: 1 mg / Kg / day.
2. Cyclophosphamide.

- SQ
- not clinical

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POLYARTERITIS NODOSA

PAN

many vs

small vs

medium vs (not large)

DEFINITION

- Vasculitis involving the small & medium-sized arteries.
- The vessels form: multiple aneurysms & multiple nodules.

weakened wall

Central area of necrosis surrounded by inflamm cells
Fibrosis

ETIOLOGY

- Uncommon, Unknown: probably autoimmune.
- May be related to INFECTION: especially HBV & HCV.. (extrahepatic)

CLINICAL PICTURE

♂ old

لوجال واحد PAN في مرقع C

- General manifestations: fever & general constitutional manifestations.
- Renal:
 - Hypertension: Due to vasculitis. → EAO → ischemia → renin ↑
 - GN: Nephritic syndrome, nephrotic syndrome, ARF, CRF, proteinuria.
- Neurological: PN, subarachnoid hemorrhage, retinal hemorrhage, blindness.
- CVS: CAD, pericarditis. (rupture of microaneurysm) Also mediated
- Pulmonary: Bronchial asthma with eosinophilia (not in classic PAN).
- GIT: Abdominal pain, hematochezia.
- Musculoskeletal: Arthralgia, Myalgia. (not common arthritis)
- Skin: Raynaud's phenomenon, palpable purpura. (extra variation of RBCs)

في
المرقع
♀ young

GN
due to
SLE
S.D.
PAN

oral
SP type
B. A. eosinophilia

Chung & Strauss
Wegener's granulomatosis
GN URTE LRTI

INVESTIGATIONS

- CBC: leucocytosis, eosinophilia, markedly elevated ESR.
- Urine: proteinuria, hematuria, casts.
- ANGIOGRAPHY: multiple microaneurysms (diagnostic).
- Biopsy: renal, muscle, skin.
- ANCA: may be positive in some cases. (specific not sensitive)

Anti-neutrophilic cytoplasmic Ab

TREATMENT

- Prednisone: 1 mg / Kg / day.
- Cyclophosphamide.

لوجال واحد
لوجال واحد
لوجال واحد

Collagen
disease
Raynaud's
Scleroderma

clinical $\leftarrow$ short long 10

RHEUMATOID ARTHRITIS

DEFINITION

articular
extra-articular
- RA is a common, chronic, systemic, inflammatory disease producing:

- Articular involvement: Symmetrical inflammatory **EROSIVE POLYARTHRITIS**. Progressive joint damage causing severe **DISABILITY**.
- Extra-articular involvement: e.g. in the lungs & many other organs.

= destruction deformity disability

INCIDENCE

- RA affects about: 2% of the population. Common
- Female to male ratio is: 3 to 1. 30 $\rightarrow$ 40y

D.D. from SLE
"Non Erosive"
10:1

ETIOLOGY

- It is still UNKNOWN.
- Factors suggested are:

autoimm + genetic

1. **Autoimmune:**
 - ① • Autoantibodies are found, e.g. **RF**, which is an autoantibody against the patient's IgG.
 - ② • Association with other autoimmune diseases such as: PA & Hashimoto's thyroiditis.
 - ③ responds to cortisol & imm drugs.
2. **Genetic:**
 - ① • RA runs in families.
 - ② • RA is commonly associated with tissue type: HLA - DR4.
3. **Infectious:**
 - Viruses: EBV.
 - Bacteria: Streptococci.

RF also in SLE and sensitive but not specific

RF also in SLE and sensitive but not specific

PATHOLOGY

- RA is disease of the **SYNOVIUM**, there are 2 main pathological features:

1. Inflammation: The synovium shows signs of **chronic inflammation**. $\rightarrow$ Growth factors
2. Proliferation: The synovium then proliferates with an outgrowth over the surface of the cartilage & forms a tumour-like mass called **Pannus**. This Pannus will destroy the intra-articular & peri-articular structures causing joint **deformities** & **dysfunction**. $\rightarrow$ cartilage bone tendons lig

- **SC nodules occur having a central area of necrosis surrounded by fibrosis.** $\rightarrow$ also in lung & spleen & macrophages

R.F. as a soft nodule SC nodules RA - 2 types

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CLINICAL PICTURE

I. ARTICULAR MANIFESTATIONS

Symptoms

- Joint pain.
- MORNING STIFFNESS.
- Disability.

دوبہ صبح (Diagnostic) lasts for 1hr at least
due to per articular effect

Signs

- Swelling, hotness, redness, tenderness, + limitation of movement.

Affected joints

- Most commonly: Per. pheral small joints of hands, wrists, knees, ankles, feet.
- Less commonly: elbows, shoulders, hips.
- Least commonly: temporomandibular, sternoclavicular.

Symmetrical polyarticular

Spared joints

- Distal interphalangeal (DIP), Sacro-iliac.

Oral

(واحد مفصل و واحد کبیر)

Deformities

- Fusiform finger:
- Ulnar deviation:
- Radial deviation:
- Boutonniere deformity:
- Swan-neck deformity:
- Z-deformity of Thumb:
- Feet & ankles:
- Knees:

swelling of the PIP is one of the most common early signs of the fingers at the MCP joints.

at the wrist joints.

flexion of the PIP & hyperextension of the DIP joints.

hyperextension of the PIP & flexion of the DIP joints.

flexion of MCP & hyperextension of IP joint.

lateral deviation of the toes.

Flexion deformity, varus or valgus deformity,

Baker's cyst.
Collection of synovial fluid
escaped from destroyed knee joint
& formed cyst in popliteal fossa

II. EXTRA-ARTICULAR MANIFESTATIONS

1. General:

fever & general constitutional manifestations.

during activity of disease

2. Skin:

SC nodules (on pressure sites & extensor tendons), Palmar erythema,

palpable purpura
Vasculitic lesions = red papules
due to extravasation of RBCs

3. Eyes:

Sjogren's syndrome (dry eyes, dry mouth),

Scleritis.
due to rupture of blood in perivascular tissue

4. Cardiac:

Pericardial disease (pericarditis, pericardial effusion), Coronary vasculitis.

5. Pulmonary:

Pleural disease (pleurisy, pleural effusion), Intrapulmonary nodules, IPF,

Caplan's syndrome (nodules + pneumoconiosis), small airway disease.

= Bronchiolitis

6. Renal:

Amyloidosis (nephrotic syndrome & CRF),

Analgesic nephropathy.

interstitial nephritis (NSAID)

7. Hematological:

Anemia (ACD, iron deficiency anemia), Palpable LN, Splenomegaly.

GRF RF in RA

1. Amyloidosis (nephrotic)
2. NSAID
3. Gold & penicillamine

↑ demand (fe def)

ACD

ass. p.A. (megaloblastic)

H-A. (due to spleen)

Sideroblastic anemia (due to drugs)

RES hyperplasia

BMT depression (drugs)

White Knight have

8. Neurological:

Carpal tunnel syndrome (the most common),

Atlanto-axial subluxation (the most serious): → cervical cord compression.

9. Soft tissues surrounding joints:

bursitis, tenosynovitis.

10. Muscles:

myopathy & muscle wasting.

11. Vasculitis:

most commonly affects the small vessels causing cutaneous vasculitic lesions.

arterioles, venules, cap

VARIANTS OF RA

all are RF +ve

1. Felty's syndrome:

Chronic RA plus:

- Splenomegaly & may be hepatomegaly & LN.
- Pancytopenia.
- Hyperpigmentation & leg ulcers. (autoimmune)

RES hyperplasia

2. Sjogren's syndrome:

Chronic RA plus:

- Dry eyes: Keratoconjunctivitis sicca.
- Dry mouth: Xerostomia.

3. Caplan's syndrome:

Chronic RA plus:

- Pulmonary nodules.
- Pneumoconiosis.

INVESTIGATIONS

1. X-ray of the joints:

- Mouse-bite erosion on the surface of the affected bone.
- Porosis of the periarticular bone.
- Loss of the joint space. (earliest sign)
- Characteristic deformities. → hyperph



Yes, yes, Porosis

mouse space ↓

Acromegaly

tuft & no loss of space

2. CBC:

- Anemia.
- ESR & CRP: elevated in proportion to activity.

3. Aspiration of synovial fluid:

- Positive RF.
- Increased protein & decreased glucose.

inflammation (AFR)

consumption due to turnover

4. Serology:

- Rheumatoid factor:

- IgM autoantibody: directed against the IgG of the patient.
- Positive in: about 80 % of the cases of RA.
- May be positive in: SLE, Sarcoidosis, SBE, TB, in 5 % of normal persons.

- Antinuclear antibodies: are positive in 30 % of the cases, non-specific.

TREATMENT

AIM OF TTT:

- Symptomatic relief of pain & inflammation.
- Modification of the course of the disease.

LINES OF TTT:

I. General measures:

- Reassurance. (Psychological)
- Rest: bed rest during activity, joint rest using splints.
- Physiotherapy.

II. Medical TTT :

1. Anti-inflammatory drugs:

a) NSAIDs:

- Aspirin: 6 gm / day.
- Indomethacin: 50 mg tds.

b) Corticosteroids:

- Prednisone: 7.5 mg / day.
- Larger doses are used only in: extra-articular manifestations: e.g. pericarditis.
- Side effects include: = CIPJcusly
 - Iatrogenic Cushing's syndrome.
 - Peptic ulcer.
 - Salt & water retention & precipitation of HF.
 - Hypertension & Diabetes.
 - Osteoporosis.
 - Delayed wound healing. destroys collagen
 - Increased incidence of infection: e.g. TB.

2. Disease – Modifying drugs: (2nd aim) immuno modulatory drugs. restore imm. system

- **Action:** They affect the natural course of the disease, probably through immune mechanisms.
- **Indications:** Progressive disease, extra-articular disease, no response to NSAIDs.
- **Drugs:**

Drug	Dose	Side effects
Penicillamine	250 mg / day orally	Rash, proteinuria
Gold auro-thiomalate	50 mg / week IM	Renal damage, PN
Hydroxychloroquine	400 mg / day orally.	Retinopathy
Sulphasalazine	1 gm twice / day orally	GIT irritation

antifibrotic
Cu chelate

B.A.

Antimalaria

IBD

damage tubules

3. Immuno-suppressive drugs:

- Used only in severe resistant cases:

- Methotrexate: 10 mg / week orally.
- Azathioprine.

Megaloblastic → f.A. antagonist
hepatotoxic →

B.M. depression

Agar also! Metabolism of

III. Treatment of complications:

1. Intra-articular corticosteroids: for rupture of the knee joint.
2. Surgery for the deformities: arthroplasty, arthrodesis.

replacement

joint fixation

Diagnosis of RA (American Rheumatoid Association)

RA is diagnosed by the presence of 4 or more of the following criteria:

1. Morning stiffness: at least 1 hour.
2. Soft tissue swelling: of 3 or more joint areas.
3. Swelling of: PIP, MCP, or wrist joint.
4. Symmetric swelling of joint areas.
5. SC nodules.
6. Positive RF.
7. X-ray: erosions and / or periarticular porosis in hands or wrist.

SERONEGATIVE ARTHRITIS

sero C-46

DEFINITION

- They are diseases characterized by:

- Involvement of the sacro-iliac joints.
- Peripheral inflammatory arthropathy.
- ABSENCE OF RHEUMATOID FACTOR.

affection of joint
eno. RF +ve

TYPES

1. Ankylosing spondylitis

- Incidence:

young adults, more in males.

- Clinical Picture:

- o Back pain: affects the sacroiliac joints & the spine causing progressive stiffness.
- o Pain: in the buttocks radiating down to the back of both legs.
- o Peripheral polyarthritis may occur.
- o Iritis & Aortic regurg may be present. *Ab mediated*

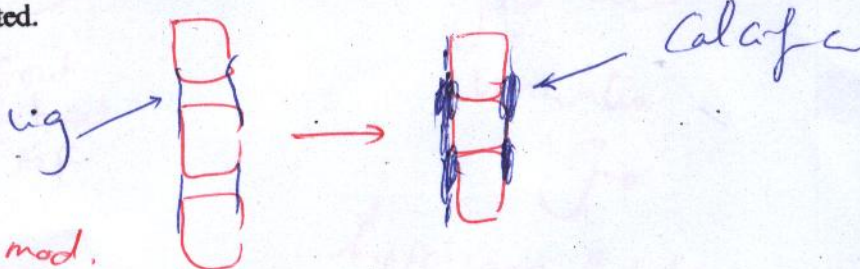
ass.

- Investigations:

- o X-ray spine: Bamboo spine: calcification of the spinal ligaments + fusion of the vertebrae.
- o ESR & CRP: are often elevated.

- Treatment:

- o NSAIDs.
- o Sulphasalazine.



2. Reiter's disease

- It consists of a triad:

seronegative arthritis, conjunctivitis, non-specific urethritis.

- It can follow:

GIT infection with Shigella or Salmonella, OR venereal infection.

- TTT:

NSAIDs.

reactively
due to release of ch mediator

3. Psoriatic arthritis

- A **seronegative arthritis** occurring in patients with psoriasis.
- A polyarthritis similar to RA but it involves the **DIP joints**.
- TTT: NSAIDs, immunosuppressive drugs.

RA spares DIP

4. Juvenile chronic arthritis

"Still's disease"

- Seronegative arthritis.
- ACUTE onset of: fever, rash, LN, hepatosplenomegaly.
- TTT: NSAIDs.

on top of chronic

5. Behcet's disease

- **RECURRENT ORAL ULCERS** (the most important feature).

- Genital ulcers.
- Uveitis.
- Vasculitis.
- Seronegative arthritis.
- Meningo-encephalitis.

- TTT: **Corticosteroids**, **Colchicines**, **Cyclosporine**.

effective

Gout
Behcet
FMF

recurrent
oral ulcers

thrombosis

venulitis

pro

hypercoagulability

clotting
factors

protein

AS

SYSTEMIC LUPUS ERYTHEMATOSUS

SLE

DEFINITION

- It is an **INFLAMMATORY, MULTISYSTEM disorder** with:

- Arthralgia & rash as the most common features.
- Cerebral & renal disease as the most serious problems.

90% ypt. (10% free)

ETIOLOGY

- It is still **UNKNOWN**.

- Factors suggested are:

1. Autoimmune:

- Autoantibodies are found, e.g. ANA, and other autoantibodies.
- Association with other autoimmune diseases such as: PA & Hashimoto's thyroiditis.
- Presence of immune complexes in the circulation leading to tissue destruction.

2. Genetic:

- SLE runs in families.
- SLE is commonly associated with tissue type: HLA - DR3.

3. Infectious:

- Viruses.
- Bacteria.

4. Others:

- Physical: Sunlight & ultraviolet light.
- Drugs: Hydralazine, procainamide, methyl-dopa, INH.
- Hormones: CCPs, estrogens, pregnancy exacerbates SLE.

PATHOLOGY

1. Vasculitis: widespread vasculitis affecting the capillaries, arterioles, venules.
2. Synovitis: inflammation of the synovium, BUT **with no erosion**.
3. Polyserositis: affection of all the serous membranes: pleura, pericardium, peritoneum.

INCIDENCE

- Sex: female to male incidence is 10 to 1.
- Age: 15 - 40 years.

- SQ
- LQ
- case

short long clinical

♀ young 20-40

rash
- arthralgia
- GN

- investg 3+ve

Abs directly

Complex

oral

ناب اللى كسر

also SBE

also PPT factors

لو كانت مفرطة الحساسية لكانت

↓ consumed hypocomplementemia

collagenase في الجلد
No Pannus

CLINICAL PICTURE

1. General:

- Fever & general constitutional manifestations.

2. Musculoskeletal:

- Arthralgia: the most common. (no limit of movt)
- Arthritis: less common.
- Myalgia, myopathy.

3. Skin:

- Malar rash: red erythema in the butterfly area.
- Photosensitivity.
- Oral ulcers.
- Alopecia: from mild loss up to total loss
- Vitiligo.
- Raynaud's phenomenon.
- Discoid lupus: butterfly rash with scaling, scarring, hyper or hypopigmentation.

4. Cardiac:

- Pericardial: pericarditis, effusion.. exudate
- Myocardial: myocarditis, cardiomyopathy. (DCM)
- Endocardial: non-bacterial endocarditis (Libman-sacks endocarditis).
- Blood vessels: CAD, thrombosis (venous, arterial).
- HYPERTENSION: due to Vasculitis or due to GN.

5. Pulmonary:

- Pleural: pleurisy, effusion.
- Lung:
 - IPF. Ab → Alveolitis (non infective pneumonia)
 - Pneumonitis.
 - Pulmonary infiltrates.
 - Pulmonary hypertension & cor pulmonale.
 - Pulmonary embolism: from DVT.

6. Neurological:

- Psychiatric: depression, psychosis.
- CEREBRAL: convulsions, stroke, aseptic meningitis, cranial nerve palsies, ataxia.
- Peripheral neuropathy.

7. GIT:

- Abdominal pain: due to peritonitis or mesenteric vasculitis.
- Anorexia, nausea, vomiting, epigastric pain.
- Hepatomegaly.

RES
due to autoimmune
GG 202

due to non-infective
gastritis by Ab
or by cortisol

White Knight Love

8. Renal:

- Pathology:

- GN of different types.
- Interstitial nephritis.

tubules ~ 15% = tubulointerstitial

- Clinical presentation:

- Nephrotic syndrome.
- Nephritic syndrome.
- CRF.
- ARF.
- Urinary abnormalities: proteinuria, hematuria.

9. Hematological:

- Cytopenias:

- Anemia: may be autoimmune hemolytic anemia
- Leukopenia.
- Thrombocytopenia.
- PANCYTOPENIA.

IGRF why SLE → anemia

the Coombs (AIHA).

oral corticosteroids
long term corticosteroids
poly cythemia jaw
mouth sores

SLE

oral

or methyl daps

- Circulating anticoagulants:

- Lupus anticoagulant: ant. cardiolip. presenting with recurrent thrombosis.
- Anti-factor VIII antibodies: presenting with Hemophilia-like picture.

- Splenomegaly.

- Lymphadenopathy.

- Impaired immunity:

due to leukopenia.

mostly in placenta
attack clotty factors
⇒ Pathological activation of
Clotty Factor

INVESTIGATIONS

1. Serology:

- Anti-ds DNA antibodies: "the most specific".
- Anti-Sm (Smith) antibodies: "the most specific".
- Antinuclear antibodies (ANA): "positive in almost all cases".
- LE cells: leucocytes with red material (DNA - anti-DNA complex) + eccentric nucleus.
- RF: may be positive in 15 % of the cases.
- Coomb's test: may be positive in AIHA.

v. specific
good +ve
sensitive
good -ve

small nuclear ptn
of RNA

Anti-VIII

Bleeding

↓ platelets

لوايته اخلف
لوم اجهه استجيب

عنه ما جيتير
Anti phospholipids

2. CBC:

- Anemia.
- Leukopenia.
- Thrombocytopenia.
- Pancytopenia.
- ESR: markedly elevated.

DNA
مخرج له
DNA - Anti-DNA cplx

1ry 2ry (SLE)
(LA → recurrent thrombosis)
oral
recurrent thrombosis
White Knight have

3. Urine analysis:

- Proteinuria.
- Hematuria.
- Casts.

} GN

4. Renal functions:

- Urea, creatinine, creatinine clearance: may be impaired.

5. Renal biopsy:

- GN of different types.
- Interstitial nephritis.

DIFFERENTIAL DIAGNOSIS

- From other causes of: acute & chronic arthropathy.
- From other causes of: polyserositis.
- From other causes of: PUO. = pyrexia of unknown origin

TREATMENT

General Medical plasmapheresis

I. General measures:

1. Avoid exposure to sunlight & ultraviolet rays.
2. Avoid drugs that exacerbate SLE. eg: INH, hydralazine, procainamide

II. Medical TTT:

1. NSAIDs:

Indications: fever, arthritis, serositis.

Drugs:

- Aspirin: 6 gm / day.
- Indomethacin: 50 mg tds.

2. Hydroxychloroquine:

Indications: skin lesions & arthralgia.

Dose: 400 mg / day orally.

3. Corticosteroids:

Indications: major organ SLE, e.g. nephritis, cerebritis, carditis, pneumonitis.

Dose:

- Prednisone: 1 mg / Kg / day orally.
- Pulse therapy: methyl prednisolone 1000 mg / day for 4 days in severe cases.

III. Plasmapheresis:

- In severe cases with nephritis or cerebritis.

& repeated if no response.

+ failure of pulse therapy

Diagnosis of SLE

SLE is diagnosed by the presence of 4 or more of the following criteria:

1. Malar rash.
2. Photosensitivity.
3. Oral ulcers.
4. Discoid lupus.
5. Arthritis.
6. Serositis.
7. Renal manifestations: persistent proteinuria, casts.
8. Neurological manifestations: psychosis, convulsions.
9. Hematological: hemolytic anemia, leucopenia.
10. Antibodies: Anti-ds DNA antibodies, Anti-Sm (Smith) antibodies, ANA.

SARCOIDOSIS

- A multisystem GRANULOMATOUS disorder of unknown etiology presenting with:

- Bilateral hilar LN
- Pulmonary infiltrations & IPF.
- Generalized LN (also epitrochlear).
- HSM, Cardiomyopathy, Arthralgias.
- Skin lesions: e.g. erythema nodosum.
- Eye lesions: bilateral iridocyclitis.

- Investigations:

1. Chest X ray: Bilateral hilar LN, pulmonary infiltrations, IPF.
2. Blood: elevated serum Ca.
3. LN biopsy: Tubercles formed of epithelioid cells, giant cells, lymphocytes, fibrosis, BUT: without caseation.
4. Kveim test: positive.
5. Serum ACE: elevated.

- Treatment:

- Corticosteroids: prednisolone 1 mg / Kg / day.

TB
Br. ca
Leukia
lyplm

Sarcoid
Strept
Sulpha
TB

oral
good
not good

لولة
لا

Ht disease
+ Ht y ↑ Ca

MM
Sarcoid
Addison
White Knight Love

SYSTEMIC SCLEROSIS

SCLERODERMA

DEFINITION

- It is an **INFLAMMATORY, MULTISYSTEM** disorder with:

- Cutaneous manifestations as the most common feature.
- **Extra-cutaneous** manifestations also occur.

ETIOLOGY

- It is still **UNKNOWN**.

- Factors suggested are:

- **Autoimmune:**

- Autoantibodies are found, e.g. **Anti-scleroderma 70**, and other autoantibodies.
- Association with other autoimmune diseases such as: PA & Hashimoto's thyroiditis.

PATHOLOGY

- There is: excessive **scarring (fibrosis)** & **vascular obliteration** of the small arteries.

Raynaud's

INCIDENCE

- Sex: Female to male ratio is: 3 to 1.
- Age: 20 – 50 years.

CLINICAL PICTURE

I. CUTANEOUS MANIFESTATIONS

- The skin is: **TIGHT**, *due to fibrous tissue by chronic infl. by Abs*, adherent to the underlying structures.
- Later on: **pigmentation, ulceration, calcification.**

1. Limited cutaneous scleroderma:

- Peripheral skin affection: *mask* hands, feet, face.
- Fingers: shiny, limited mobility.
- Face: *mask face, fish mouth.*

2. Diffuse cutaneous scleroderma:

- Diffuse skin affection: trunk, hands, feet, **face**.



II. EXTRA-CUTANEOUS MANIFESTATIONS

1. GIT:

- **Mouth:** fish mouth.
- **Oesophagus:** loss of peristalsis, dilatation, dysphagia, **GORD**.
↓ motility
- **Small intestine:** malabsorption.
- **Large intestine:** constipation.

2. CVS:

- **Pericardial:** pericarditis, effusion.
- **Myocardial:** **restrictive cardiomyopathy**.
- **Endocardial:** MR, TR.
- **Blood vessels:** **RAYNAUD'S PHENOMENON** (very common).
- **Hypertension:** due to renal vasculitis.

3. PULMONARY:

- **Chest wall:** hard skin leading to: **Restrictive hypoventilation**.
- **Pleura:** pleurisy.
- **Lung:** **IPF**, aspiration pneumonia (due to GORD).
- **Blood vessels:** pulmonary hypertension & cor pulmonale.

4. RENAL:

- **Malignant hypertension:** due to obliterative endarteritis of the renal vessels. *EAO*
- **CRF.**

5. MUSCULOSKELETAL:

- **Arthralgia:** the most common.
- **Arthritis:** less common.
- **Myalgia,** myopathy.

Crest Syndrome

C: calcinosis

R: Raynauds

E: esophageal dysmotility

S: sclerodactyly (hand fingers)

T: Telangiectasia Dil B.v.

INVESTIGATIONS

1. Serology:

- **Anti - Scleroderma 70 antibodies:** "the most specific"
- **ANA, RF, LE cells:** "not specific"

2. CBC:

- **Anemia.**
- **Markedly elevated ESR.**

3. Imaging:

- **Ba swallow:** Dilated oesophagus.
- **Chest X-ray:** IPF.

TREATMENT

1. Specific ttt:

- Corticosteroids: Prednisone, 1 mg / Kg / day.
- Immunosuppressives: Cyclophosphamide.
- Antifibrotics: Penicillamine. *antifibrotic*

2. Symptomatic ttt:

- e.g. For Raynaud's phenomenon: Calcium channel blockers, alpha blockers.

eg Gerd PPI omeprazole

B.V. ليفتو

DERMATOMYOSITIS & POLYMYOSITIS

1. Skin manifestations:

- Erythematous rash.

MS & SK

MS

PMs Br. Ca.

2. Muscle manifestations:

- Muscle pain & tenderness.
- Features of myopathy.

3. Malignancy:

- Present in 18 % of the cases: e.g. bronchial carcinoma.

Oral

OVERLAP SYNDROME

- Patients with features of more than one collagen disease.

Mixed C.T. Disease.
MCTD

- Patients with features of:

SLE,

Scleroderma,

Polymyositis.

overlap

(1)

(2)

(3)

AMYLOIDOSIS

DEFINITION

- Abnormal deposition of amyloid proteins in the tissues.

TYPES

1. PRIMARY:

- Heart:
- GIT:
- Neurological:
- Joints:

"Hereditary"

- Cardiomyopathy.
- Macroglossia, Malabsorption.
- PN, Carpal tunnel syndrome.
- Arthritis.

DCM
RCH

obs. of lacteals
(small int. Caused)

2. SECONDARY:

- It is due to:
- It presents with:

Chronic suppurative disease (SLS), TB, RA, FMF.
HSM, Nephrotic syndrome.

infectn & inflammation

supp lung syndrome

INVESTIGATIONS

1. Investigations of the cause & the complications.
2. Rectal biopsy: amyloid material by "Congo red stain".

diagnostic

TREATMENT

- Treatment of the cause & the complications.

TB: bacteriology nephrotic: urine for ph $> 3g / 24 \text{ hour}$

Blood malignancy

MULTIPLE MYELOMA

- Malignant disorder of plasma cells in OLD AGE presenting with:

1. Skeletal: Bony pains, pathological fracture, skull osteolytic lesions, $\uparrow \text{Ca}$
2. Renal: RENAL FAILURE & Bence-Jones proteinuria.
3. Neurological: Focal spinal cord compression.
4. Blood: ANEMIA (BM infiltration, BM inhibition, uremia).
5. Bleeding: coating of platelets & clotting factors by abnormal proteins.
6. Hyperviscosity.

- Investigations:

1. Skeletal X-ray: Multiple osteolytic lesions in the bones.
2. Renal: Impaired renal functions, Bence-Jones proteinuria.
3. Blood: Anemia, elevated ESR, elevates serum Ca.
4. BM exam: Full of plasma cells.
5. PLASMA PROTEIN ELECTROPHORESIS: Monoclonal hypergammaglobulinemia.

- Treatment:

- Chemotherapy, Radiotherapy, BMT.

BMT replaced by
malignant plasma cells

M A G E D

to see if it's really there

RF

$\uparrow \text{Ca}$ $\uparrow$ protein
nephrocalcinosis

high amino
ketones
2000
500
45
M.M.
"bi"

oligo clonal
- Hs

including catalsal

thy disease

total

$\downarrow \text{Ca}$